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Sleep disordered breathing and subclinical impairment of respiratory function are common in sporadic inclusion body myositis

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Abstract

Relatively little is known about frequency and extent of respiratory problems in sporadic inclusion body myositis (IBM). To address this issue a study of peripheral muscle and respiratory function and related symptoms was performed in a cohort with biopsy-proven IBM. Dyspnoea, daytime sleepiness, dysphagia, spirometry, respiratory muscle strength, arterial blood gas tensions and ventilation during sleep were assessed. Sixteen patients were studied (10 males; age 68.1 ± 9.9 years; disease duration 11.9 ± 5.0 years; body mass index 28.5 ± 4.0 kg/m^2). Four reported excessive daytime sleepiness; 8 had at least mild dysphagia; forced vital capacity was <80% predicted normal in 7; sniff nasal inspiratory pressure was reduced in 3; daytime hypoxemia was present in 9 and hypercapnia in one. Sleep study was performed in 15 and revealed sleep disordered breathing (apnoea–hypopnoea index 23.4 ± 12.8 (range 7–50.3) events/h) in all. There were no consistent relationships between respiratory function impairment, occurrence of sleep disordered breathing, and severity of peripheral muscle weakness. Thus, asymptomatic impairment of respiratory function was common and sleep disordered breathing observed in all patients tested, irrespective of daytime respiratory function. This suggests respiratory function testing, including sleep study, should be performed routinely in IBM, irrespective of peripheral muscle function or other disease severity parameters.

Keywords: Sporadic inclusion body myositis; Respiratory function; Obstructive sleep apnea; Sleep disordered breathing; Respiratory failure

1. Introduction

The idiopathic inflammatory myopathies are a heterogeneous group of disorders which differ in their clinical, immunopathological features and diagnostic criteria [1]. The three most common forms are dermatomyositis, polymyositis, and inclusion body myositis (IBM) which is the most common acquired muscle disease in older individuals [2]. Several studies

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have addressed the occurrence of respiratory complications in dermatomyositis and polymyositis, revealing a high prevalence of problems, including interstitial lung disease, hypercapnic respiratory failure due to muscle weakness, and obstructive sleep apnoea [3,4]. Much less is known about respiratory problems in IBM but case reports of respiratory failure in IBM patients support the hypothesis of possible respiratory muscle involvement [5–8]. However, these reports provide no guidance regarding the prevalence of respiratory problems in IBM. Furthermore they have used a variety of measures of wakeful respiratory function, with no attempt to relate these to peripheral muscle function or to breathing during sleep. These are significant deficiencies, as muscles are not uniformly affected in IBM, and where respiratory muscles or pharyngeal muscles are involved, sleep can be a vulnerable state because of the risk of sleep related hypoventilation or upper airway obstruction (i.e. obstructive sleep apnoea (OSA)). Sleep-related reduction in ventilatory drive is associated with nocturnal hypoventilation in patients with moderate respiratory muscle weakness who may not yet have developed wakeful respiratory failure. Thus, sleep hypoventilation can be a harbinger of respiratory failure in neuromuscular disorders in which the respiratory muscles are involved [9]. It is known that OSA is common in disorders associated with pharyngeal muscle weakness [10]. IBM is such a condition and a recent study suggests that it can be associated with sleep disordered breathing, including OSA [11].

To better characterise these issues we used a standard array of tests to measure wakeful respiratory function, as well as ventilation during sleep, in a cohort of 16 patients with proven inclusion body myositis. Our purpose was to determine the frequency of respiratory muscle weakness, how this is related to peripheral muscle strength, and its impact on wakeful and sleep-related breathing function. We hypothesised that deficits would be found and that the impacts would be more sensitively reflected in sleep-related than wakeful measures of breathing function.

2. Materials and methods

2.1. Subjects

A cross-sectional study was performed of all consenting biopsy-proven IBM patients currently attending the clinics at the Australian Neuromuscular Research Institute (Perth, Western Australia). Sixteen patients who fulfilled the Griggs et al. diagnostic criteria [12] for definite IBM were included (10 male; mean($\pm$SD) age 68.1 $\pm$ 9.9 years). Current smokers were excluded. None of the patients used oxygen therapy. Ten patients were on immunosuppressive treatment (5 prednisolone + methotrexate; 2 prednisolone + mycophenolate; 2 methotrexate; 1 azathioprine). The study protocol was approved by the Sir Charles Gairdner Hospital Human Research Ethics Committee.

2.2. Measurements

Anthropometric data (age, gender, height, weight) were collected. Disease severity was assessed by the IBM functional rating scale (IBM-FRS) [13]. This scale assesses function across 10 activities of daily living scoring each from 0 (unable to perform) to 4 (normal). The closer the total score is to the maximum possible of 40 the better the functional status of the patient. The strength in the quadriceps and other upper and lower limb muscles was evaluated by manual muscle testing (MMT) using a 0–10 point modification of the Medical Research Council (MRC) scale for muscle strength, which grades it from 0 (no movement detected) to 10 (normal) [14]. The degree of breathlessness and its relationship to activity was assessed using the MRC Dyspnoea Scale which is a 5 point scale from 1 (breathless only with strenuous exercise) to 5 (too breathless to leave house) [15]. The degree of daytime sleepiness was assessed using the Epworth Sleepiness Scale (ESS), which questions likelihood of dozing from 0 (no chance) to 3 (high chance) under 8 different situations [16]. The closer to the maximum score of 24 the greater the degree of daytime sleepiness. The presence and severity of dysphagia was quantified using the Dysphagia Outcome and Severity Scale (DOSS), a 7 level scale from 1 (severe dysphagia) to 7 (normal in all situations) [17].

Spirometry (FEV1, FVC and FEV1/FVC) was performed (EasyOne® Plus Spirometer, NDD Medical Technologies Inc., Andover, MA) in both sitting and supine positions according to the standards of the American Thoracic Society [18]. Inspiratory muscle strength was evaluated using sniff nasal inspiratory pressure (SNIP) (MicroRPM, Carefusion Corporation, San Diego, CA). SNIP was measured according to the European Respiratory Society guidelines [19,20].

Gas exchange was assessed using arterialised capillary blood from the earlobe. Patients were all breathing room air (without oxygen supplementation) at the time of measurement. The samples were analysed (ABL700, Radiometer Medical ApS, Copenhagen, Denmark) for HCO_3 concentration and partial pressures of oxygen (PaO_2) and carbon dioxide ($PaCO_2$).

Ventilation was studied during sleep using a dual channel (oximetry, nasal airflow) portable home monitoring device (ApneaLink™, Resmed Ltd., Bella Vista, Australia). The data were uploaded in European data format (EDF) into a laboratory polysomnographic analysis system (E series, Compumedics Ltd., Melbourne, Australia) for subsequent manual analysis to define: apnoea–hypopnoea index (AHI); arterial oxygen saturation (SpO_2); oxygen desaturation index for number of desaturation events of 3% or more per hour of

estimated sleep time (ODI-3%); and proportion of total sleep time spent at or below a saturation of 90% (TST $\leqslant$ 90). AHI was defined as the number of apnoeas (reductions in amplitude of nasal airflow to <10% of baseline for $\geqslant$10 s) and hypopnoeas (>50% reduction in nasal airflow from baseline (but not to a degree classifiable as an apnoea) or a lesser reduction in airflow associated with >3% oxygen desaturation with events lasting $\geqslant$10 s) per hour of estimated sleep time [21].

Statistical analysis was performed with SPSS v15.0 software (IBM, Armonk, NY). Interrelationships between ventilatory capacity, gas exchange and polysomnographic parameters were examined using Pearson correlation coefficients and simple regression linear analysis. The Mann–Whitney *U* Test was used to compare the presence of sleep apnoea/hypopnoea events in patients with and without dysphagia and to assess differences between patients with and without immunosuppressive treatment. All results are presented as mean ± standard deviation. A *p*-value of <0.05 was considered as significant.

3. Results

Patient demographics, clinical data and results of measures of wakeful respiratory function, sleep breathing parameters and daytime capillary blood gas tensions are summarised in Table 1. Sixteen patients were studied. They were all the eligible patients registered at our clinic at commencement of data collection in February 2013. Complete data were collected on all patients apart from: (a) absent sleep data in patient 12; and (b) absent blood gas data and SNIP data in patient 13, because of logistical difficulties in each case.

3.1. Wakeful respiratory function

None of the 16 subjects had any previously identified wakeful respiratory problems, apart from one with a history of asthma (subject 2). She and two others (subjects 9 and 11) had an FEV1/FVC ratio <70%, suggestive of airflow obstruction. One of these (subject 9) had a previous 20-pack year smoking history. Six had a restrictive spirometric pattern (FVC < 75% predicted value with normal or elevated FVC/FEV1 ratio). None had evidence of paradoxical abdominal wall movement in the supine position, and only one (subject 10) had a fall in vital capacity of >8% with change from erect (seated) to supine posture.

The SNIP was substantially reduced (<60 cm H_2O) in 3 subjects (subjects 1, 3 and 4), indicative of inspiratory muscle weakness. All these subjects had a restrictive ventilatory defect. The subject with the lowest SNIP (subject 1) also had one of the lowest FVC and was the only one who was in type 2 (hypercapnic) respiratory failure ($PaCO_2 > 6$ kPa). Breathlessness was not prominent in any of the subjects. No subject had an MRC dyspnoea score >2; half the subjects had a score of 1 and half of 2.

3.2. Wakeful blood gases

Wakeful capillary blood gases demonstrated hypoxemia ($PaO_2 < 10.5$ kPa) in 9 patients and hypercapnia ($PaCO_2 > 6$ kPa) in one. While there were no significant correlations between blood gas parameters (PaO_2, $PaCO_2$, HCO_3) and ventilatory capacity (FVC, FEV1), the patient with the lowest SNIP and near lowest FVC (subject 1) was the only one with hypercapnia.

3.3. Ventilation during sleep

Five patients (subjects 3, 4, 13, 15 and 16) reported regular snoring and two of them had previously diagnosed OSA. Four reported excessive daytime sleepiness (ESS $\geqslant$ 10); mean ESS was 5.7 ± 4.1 (range 0–12). None reported morning headache.

Sleep studies were performed in 15 patients. During sleep, 10 patients demonstrated significant sleep hypoxemia (TST $\leqslant$ 90 of $\geqslant$2%), with 6 having a nadir SpO2 below 85%. Nocturnal desaturation episodes were very frequent (ODI-3% 18.2 ± 8.4 events/h). The mean AHI was 23.4 ± 12.8 events/h. All patients had sleep apnoea (AHI $\geqslant$ 5 events/h), but with different severity grades: mild (AHI 5–15 events/h) in 5, moderate (AHI 15–30 events/h) in 6 and severe (AHI > 30 events/h) in 4. ODI and AHI were highly correlated ($r = 0.9$, $p < 0.01$). Daytime PaO_2 was significantly negatively correlated with the TST $\leqslant$ 90 ($r = -0.55$, $p = 0.43$). We found no significant correlation between sleep oxygenation (TST $\leqslant$ 90) and ventilatory capacity or BMI. The AHI was not related to ventilatory capacity, gas exchange parameters or BMI.

The ESS score was significantly correlated with ODI ($r = 0.66$, $p = 0.01$), AHI ($r = 0.63$, $p = 0.01$) and negatively correlated with FVC ($r = -0.62$, $p = 0.01$) and FEV1 ($r = -0.58$, $p = 0.02$).

3.4. IBM functional rating scale, quadriceps strength and other parameters

Functional status (IBM-FRS) varied considerably between subjects. There were no correlations between disease duration and the IBM-FRS score, vital capacity, parameters of gas exchange or sleep ventilation, but quadriceps muscle strength was negatively correlated with disease duration ($r = -0.70$, $p = 0.002$). In terms of disease severity, the only correlations found were negative relationships between SNIP and both IBM-FRS ($r = -0.66$, $p = 0.008$) and quadriceps muscle strength ($r = -0.49$, $p = 0.06$) and a negative correlation between FEV1 and quadriceps muscle strength ($r = -0.53$, $p = 0.04$). We found no difference in results for the variables studied between patients with and without immunosuppressive treatment.

Table 1
Demographic, clinical, respiratory and sleep data from the 16 subjects.

Subject	Demographics				Clinical scales					CK	Respiratory function				Sleep parameters			Arterial blood gases		
	Age (years)	Sex	BMI (kg/M^2)	Disease duration (years)	MRC dyspnoea	ESS	DOSS	MMT	IBM-FRS	CK (IU/L)	SNIP (cm H_2O)	FVC erect (%pred)	FEV_1/FVC Erect (%)	ΔFVC erect-supine (%)	AHI (/hr)	Mean SaO_2 (%)	TST ⩽ 90 (%)	$PaCO_2$ (kPa)	PaO_2 (kPa)	HCO_3 (mmol/L)
1	68	F	30.1	8	2	8	3	7.5	28	179	47	61	81.9	0	35.7	94	5	6.6	9.25	30
2	63	F	32	9	2	7	7	6.5	26	1262	81	70	60.7	5	28.7	90	20	5.5	8.8	27
3	76	F	25.5	14	2	7	7	4.5	37	36	58	57	87.4	0	31.6	93	0	4.9	10.0	24.1
4	79	M	28.1	4	2	4	7	6.5	32	163	57	73	73	2	20	93	0	4.0	15.2	21.2
5	74	M	32.7	6	1	10	5	8	19	238	79	66	74.9	0	12.6	93	2	5.1	11.7	23.4
6	72	F	25.8	14	1	3	7	2	18	170	96	89	74.6	7	8.1	96	0	5.0	9.75	25.1
7	71	M	32.7	14	1	12	7	2.5	26	399	76	73	73.9	0	50.3	94	3	4.4	11.1	23.2
8	64	M	36.9	16	2	1	5	3.5	21	132	88	97	78.9	0	20.9	94	2	3.4	11.8	16.8
9	67	F	24.1	6	2	1	4.5	9	29	75	70	104	67	3	14.3	92	10	4.8	8.5	23.6
10	73	M	29.6	17	1	12	4.5	2	25	56	92	89	77.1	9	41.3	93	6	5.7	9.3	26
11	78	M	27.7	7	1	6	7	2	31	191	91	97	61.6	2	15.9	92	6	4.75	10.85	23.6
12	66	F	24.5	11.5	2	5	3	5	33	112	60	90	78.3	0	–	–	–	4.8	9.1	24.6
13	55	M	32	13	2	11	7	8	32	272	–	73	71.8	1	26.2	92	5.3	–	–	–
14	40	M	25.7	12	1	2	6	4.5	26	220	73	97	80	0	7	95	0	5.2	10.1	24.6
15	65	M	22.1	18	1	2	4	2	27	331	81	91	81	0	9.5	94	1	5.6	10.6	26.2
16	78	M	26.9	22	1	0	4	1.5	32	666	64	91	73	0	28.6	91	9	5.5	7.9	25.7
Mean (SD)	68.1 (9.9)		28.5 (4.0)	11.9 (5.0)	1.5 (0.5)	5.7 (4.1)	5.5 (1.5)	4.7 (2.6)	27.6 (5.2)	281 (306)	74.2 (14.6)	82.5 (14.5)	74.7 (7.1)	1.8 (2.8)	23.4 (12.8)	93.1 (1.5)	4.6 (5.4)	5.02 (0.76)	10.26 (1.78)	24.3 (2.9)

BMI = body mass index; MRC dyspnoea = Medical Research Council Dyspnoea Scale; ESS = Epworth Sleepiness Scale; DOSS = Dysphagia Outcome and Severity Scale; MMT = manual muscle testing score for quadriceps strength on 10-point scale (average of right and left sides); IBM-FRS = inclusion body myositis functional rating scale (maximum score 40); CK = creatine kinase; SNIP = sniff inspiratory pressure; AHI = apnoea–hypopnoea index; TST ⩽ 90 = % total sleep time spent at or below a $SpO_2 < 90\%$. For further explanations see text.

3.5. Swallowing function

Half of the subjects had at least mild impairment of swallowing (DOSS ⩽ 5) and two had videofluoroscopically confirmed swallowing dysfunction. Mean DOSS was 5.5 ± 1.5 (range 3–7). There was no significant relationship between DOSS and any of the parameters of wakeful or sleep respiratory function, including AHI.

4. Discussion

This study of an unselected cohort of patients with sporadic IBM identified a range of respiratory problems of varying severity, from trivial to highly clinically significant. Although respiratory symptoms were not prominent in the group as a whole, one of the 16 patients was in overt type 2 respiratory failure and 7 had vital capacities of less than 75% predicted normal, suggesting that they had significant ventilatory impairment. In many cases subtlety or absence of daytime symptoms made such issues difficult to identify clinically. Of particular note was the finding of sleep disordered breathing in all 15 patients in whom sleep studies were performed.

The high occurrence of sleep disordered breathing in this cohort is consistent with findings of a recent Italian study [11]. In that study a polysomnographic evaluation was used and demonstrated sleep disordered breathing in 7 of 12 patients in whom adequate data were obtained. While a lower proportion of sleep disordered breathing than observed in our patients it was notable that in 6 of these 7 the events were predominantly obstructive; the seventh had a tracheotomy but had mild central sleep apnoea.

The dual channel device used to assess breathing during sleep in the present study is limited in its capacity to distinguish obstructive (reduced flow, ongoing effort) from central (concordant reduction in flow and effort) breathing disturbances by its lack of an effort sensor. Nevertheless there were several features evident on breath-by-breath analysis of the raw flow data recorded by the device that suggest sleep disordered breathing was predominantly obstructive in each of our subjects. Flow limitation (flattening of the inspiratory flow profile) and/or snoring (detected from observation of a characteristic high frequency component of the inspiratory flow signal) were commonly observed in association with respiratory events in each subject. The snoring was sometimes observed during development of events and often during recovery breaths following them. The morphology of the flow changes observed during a substantial majority of events also favoured obstruction with a progressive decrease in flow (to cessation in the case of apnoeas) followed by an abrupt resumption of flow on termination of the event. This was in contrast to waxing/waning flow patterns typically seen in periodic breathing or to prolonged periods of reduced respiratory flow (without flow limitation) seen in sleep hypoventilation.

Sleep is associated with reduced skeletal muscle activation and ventilatory drive, physiological changes that make it a vulnerable state for patients with weak pharyngeal or respiratory muscles. Deficiencies in these muscles may become manifest during sleep as upper airway obstruction or hypoventilation, even if such

impairments are insufficient to cause wakeful symptoms [9]. OSA can be relatively asymptomatic, particularly in its milder forms. Daytime sleepiness, a common symptom resulting from associated sleep disruption, is inconsistently present. Nevertheless, whether overtly symptomatic or not, OSA is associated with a range of co-morbidities that make it important to identify and treat [22].

A high occurrence of OSA in IBM might be expected given the frequent presence of overt or covert pharyngeal muscle weakness in these patients [2]. Although half of the patients in the present study also had dysphagia, the lack of a direct relationship between the DOSS and sleep disordered breathing severity suggests that wakeful pharyngeal muscle dysfunction is often subclinical and that the DOSS is insensitive to its presence. While 50% of the cohort had a DOSS of 5 or less, indicating at least moderate swallowing impairment, the findings of this study suggest that *sleep-related* pharyngeal muscle dysfunction, resulting in OSA, could be present in more, a possibility that should be considered in all patients with IBM.

Apart from pharyngeal muscle dysfunction, impaired respiratory muscle function was also commonly observed in this patient group. While breathlessness was not a prominent feature in any of the patients, vital capacity was reduced (<75% of predicted normal) in 7 of them. In one (subject 2), this could be attributed to smoking related airflow obstruction, as evidenced by her reduced FEV1/FVC ratio. In the remaining 6 patients it is likely that their low vital capacities were due to respiratory muscle weakness: in the 5 of them in whom SNIP was also measured it was found to be <80 cm H_2O in each, while in 3 of these it was <60 cm H_2O, which is below the accepted lower limit of normal for this test [23].

The finding of an inverse relationship between quadriceps muscle strength and both ventilatory capacity (FEV1) and inspiratory muscle strength, as reflected by SNIP, was unexpected and the possibility that this negative relationship may be factitious cannot be excluded, particularly given the relatively low number of patients studied. Nevertheless, it may serve to underline the heterogeneous nature of the muscle involvement in IBM and the absence of a fixed relationship between the degree of respiratory muscle weakness and limb muscle weakness. Similarly, there was a negative correlation between SNIP and IBM-FRS. These findings suggest that respiratory status of IBM patients cannot be inferred from peripheral muscle function or other non-respiratory disease severity parameters. They also raise the possibility that there may be adaptive changes in respiratory muscle function in the face of progressive weakness of the limb muscles and associated impairment of mobility and difficulty in performing activities of daily living.

Only one patient (subject 1) was in overt type 2 respiratory failure, with a wakeful $PaCO_2$ of 6.6 kPa and associated hypoxemia (9.25 kPa). In this patient the SNIP was the lowest observed in the cohort and FVC was almost the lowest, consistent with respiratory muscle weakness. Despite this, breathlessness was not a feature in this patient, illustrating the covert nature of the respiratory problems. Notably, neither this patient nor any of the others had a substantial (>10%) decrease in FVC between the upright and supine postures as is usually present with diaphragmatic weakness [24]. This suggests that any respiratory muscle weakness is largely due to weakness of rib cage and abdominal musculature rather than the diaphragm. The other notable feature in Subject 1 was her high AHI, suggesting that she had substantial pharyngeal as well as respiratory muscle weakness. Her reduced DOSS supports this contention. The combination of OSA and respiratory muscle impairment is known to be potent in the pathogenesis of wakeful respiratory failure [9].

It is noteworthy that 8 of the other 15 subjects also had mild-to-moderate daytime hypoxemia (PaO_2 from 7.9 to 10.1 kPa), but with normal $PaCO_2$, suggesting that the reduced PaO_2 is likely to be due to disordered gas exchange (e.g. from age-related changes or other factors such as obesity-related atelectasis) rather than alveolar hypoventilation. While sleep hypoventilation is a possible explanation for the finding of high TST $\leqslant$ 90 findings in some individuals, those with greatest TST $\leqslant$ 90 (subjects 2, 9 and 16) all had a wakeful PaO_2 of <9 kPa and $PaCO_2$ of $\leqslant$5.5 kPa, suggesting, in them at least, that nocturnal hypoxemia reflected disordered gas exchange that was also evident when awake. Indeed, the direct relationship we observed between daytime PaO_2 and nocturnal TST $\leqslant$ 90 may largely reflect different (wakeful and sleep-related) measures of an independently determined gas exchange abnormality (eg age related [25,26].) However, it also raises the possibility that sleep hypoxemia could be making a direct contribution to the daytime hypoxemia [27].

We found no significant differences in any of the parameters studied between patients with and without immunosuppressive treatment. This suggests that the changes in respiratory function observed in the present study cannot be simply explained by chronic utilisation of corticosteroids or other immunosuppressive drugs. Nevertheless these findings do not exclude the possibility of respiratory effects from such drugs in other patients and/or changes in other more complex respiratory assessments, such as high-resolution imaging or gas transfer factor, not assessed here.

This study has limitations. Firstly there was the relatively low number of patients enroled, which reflects the low prevalence of IBM in the general population. A survey in 2008 in Western Australia, the setting of the present study, reported a prevalence of 14.9 cases per million population [28]. Nevertheless the present cohort was unselected, representing all eligible patients attending a large clinic specialising in neuromuscular disorders. This limited the capacity to draw inferences from this

descriptive study. Secondly, the dual channel (oximetry, respiratory flow) ApneaLink™ device provides a limited assessment of sleep disordered breathing as lack of an effort sensor reduces the capacity to distinguish central from obstructive events and lack of sleep staging (as provided for by gold standard polysomnography) means events are indexed according to estimated rather than actual sleep time.

5. Conclusions

The high frequency of pharyngeal and respiratory muscle involvement in this patient cohort, and the frequently subclinical nature of these problems, suggests that they should be specifically looked for in all cases of IBM. Investigations during sleep are particularly important where potential pharyngeal or respiratory muscle impairment is concerned, as mild dysfunction may only be apparent in the presence of sleep related reductions in muscle activation and ventilatory drive. Sleep disordered breathing, which was observed in all the patients tested and was often severe, is amenable to treatment by both ensuring potential aggravating factors such as alcohol and drug consumption and weight excess are addressed and, commonly, use of positive airway pressure therapies.

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